



IN THE UNITED STATES PATENT AND TRADEMARK OFFICE

Applicants: Thomas J. Hudson, James Engert and Andrea Richter
Application No.: 09/693,205 Group: 1646
Filed: October 20, 2000 Examiner: Not yet assigned
For: Identification of ARSACS Mutations and Methods of Use Therefor

| | |
|---|--------------------------------------|
| CERTIFICATE OF MAILING | |
| I hereby certify that this correspondence is being deposited with the United States Postal Service with sufficient postage as First Class Mail in an envelope addressed to Assistant Commissioner for Patents, Washington, D.C. 20231 | |
| on <u>4/5/01</u> | <u>Paula DePeltreau</u> Signature |
| <u>PAULA DEPELTREAU</u> Typed or printed name of person signing certificate | |

REPLY TO NOTICE TO FILE MISSING PARTS OF APPLICATION

Box Missing Parts
Assistant Commissioner for Patents
Washington, D.C. 20231

Sir:

In reply to the Notice to File Missing Parts dated December 14, 2000, the following documents and fees are being submitted for filing in the captioned application. A copy of the Notice is attached.

EXECUTED DECLARATION/POWER OF ATTORNEY

(Separate transmittal letter and postcard not required)

FILING FEE - with Fee Transmittal for Patent Applications in duplicate.

(Separate transmittal letter and postcard not required)

SURCHARGE - surcharge fee of \$130.00.

(Separate transmittal letter and postcard not required)

SEQUENCE LISTING - Filed concurrently and is attached.

(Separate transmittal letter and postcard required)

TRANSMITTAL OF SUBSTITUTE DRAWINGS

Sheets of substitute drawings consisting of Figs. 1 - [] are enclosed.
(Separate transmittal letter and postcard not required)

 PETITION FOR EXTENSION OF TIME

Applicants hereby petition to extend the time to respond to the Notice to File Missing Parts dated December 14, 2000 for two months from February 14, 2001 to April 14, 2001. The appropriate fee of \$390 is included in the enclosed check.

A [] month extension of time to respond to the Notice to File Missing Parts dated [] was filed on [] with payment of a \$[] fee.

Applicant hereby petitions for an additional [] month extension of time to respond to the Notice to File Missing Parts. The appropriate fee of \$[] is included in the enclosed check.

(Separate Petition for Extension of Time and postcard not required)

 REQUEST FOR CORRECTED FILING RECEIPT - Filed concurrently and is attached.

(Separate transmittal letter and postcard required)

 STATEMENT CLAIMING SMALL ENTITY STATUS

Was filed on [].

Is enclosed herewith.
(Separate transmittal letter and postcard not required)

In view of the small entity status of the captioned application, we hereby request a reimbursement of 50% of the filing fees in the amount of \$[] which were paid on [] to be deposited in Deposit Account No. 08-0380.

The fees required for filing the indicated documents are enclosed in the form of a check in the total amount of \$2,612.00. Please charge any deficiency or credit any overpayment in the fees that may be due in this matter to Deposit Account No. 08-0380. A copy of this letter is enclosed for accounting purposes.

Respectfully submitted,

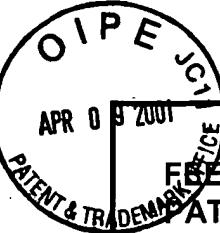
HAMILTON, BROOK, SMITH & REYNOLDS, P.C.

By Lisa M. Treannie
Lisa M. Treannie
Registration No.: 41,368
Tel.: (781) 861-6240
Fax: (781) 861-9540

Lexington, Massachusetts 02421-4799
Date:

4/5/01

APR 09 2001



| | | |
|--|------------------------|------------------|
| FEE TRANSMITTAL FOR PATENT APPLICATIONS | Attorney Docket Number | 2825.1021-003 |
| | Application Number | 09/693,205 |
| | First Named Inventor | Thomas J. Hudson |

CLAIM CALCULATION (includes any preliminary amendment)

| CLAIMS | (1) FOR | (2) NUMBER FILED | (3) NUMBER EXTRA | (4) RATE | (5) CALCULATIONS |
|--------|---|------------------|---|--------------------------------------|------------------|
| | TOTAL CLAIMS (37 CFR 1.16(c) or (j)) | 39 - 20* = | 19 | x \$ 18 = | \$ 342 |
| | INDEPENDENT CLAIMS (37 CFR 1.16(b) or (i)) | 16 - 3** = | 13 | x \$ 80 = | \$ 1040 |
| | MULTIPLE DEPENDENT CLAIMS (if applicable) (37 CFR 1.16(d)) | | | + \$ 270 = | \$ |
| | | | | BASIC FEE (37 CFR 1.16(a) or (h)) | \$ 710 |
| | | | Total of above Calculations = | | \$ 2092 |
| | | | Reduction by 50% for filing by small entity (37 CFR 1.9, 1.27, 1.28) = | | \$ |
| | | | TOTAL = | | \$ 2092 |
| | | | Surcharge - Late Filing of Declaration or Filing Fees (37 C.F.R. 1.16(e)) = | | \$ 130 |
| | | | Petition for Extension of Time Fee (37 C.F.R. 1.17) = | | \$ 390 |
| | | | Assignment Recordation Fee = (only when filed with application) | | \$ |
| | * Reissue claims in excess of 20 and over original patent ** Reissue independent claims over original patent | | TOTAL = | | \$ 2612 |

1. Small entity status: | 04/11/2001 JADD01 00000024 09693205
 | 05 FC:116 390.00 OP

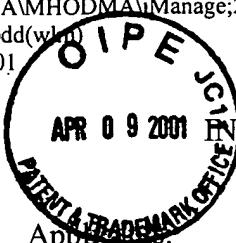
- a. A small entity statement is enclosed.
- b. A small entity statement was filed in the prior non-provisional application and such status is still proper and desired.
- c. Is no longer claimed.

2. A general authorization is hereby granted to charge deposit account number 08-0380 for any fees required under 37 CFR 1.16 and 1.17 in order to maintain pendency of this application. A copy of this authorization is enclosed for accounting purposes.

3. A check is enclosed for \$2612.00. | Please charge \$[] to Deposit Account No. 08-0380.

4. Other: _____

| | | | |
|---------------------------------------|------------------|-------------|---------------|
| Signature | Lisa M. Treannie | Date | April 5, 2001 |
| Submitted by Typed or Printed Name | Lisa M. Treannie | Reg. Number | 41,368 |



THE UNITED STATES PATENT AND TRADEMARK OFFICE

Application: Thomas J. Hudson, James Engert and Andrea Richter

Application No.: 09/693,205 Group: 1646

Filed: October 20, 2000 Examiner: Not assigned

For: Identification of ARSACS Mutations and Methods of Use Therefor

CERTIFICATE OF MAILING

I hereby certify that this correspondence is being deposited with the United States Postal Service with sufficient postage as First Class Mail in an envelope addressed to Assistant Commissioner for Patents, Washington, D.C. 20231

on 4/5/01 Paula Depeltreau
Date Signature

PAULA DEPELTREAU

Typed or printed name of person signing certificate

REQUEST FOR CORRECTED FILING RECEIPT
FOR UTILITY APPLICATION

Office of Initial Patent Examination
Customer Service Center
Assistant Commissioner for Patents
Washington, D.C. 20231

Sir:

We hereby request that the following items of information be corrected in the Filing Receipt for the subject application received in this office on December 22, 2000.

The errors and corrections appear below.

The Atty Docket No. for this case should read "2825.1021-003", not "2852.1021-003".

09/693,205

-2-

Enclosed are a copy of the Filing Receipt with changes noted in red, a copy of the Utility Patent Application Transmittal, and a copy of page 1 of the Application as filed on October 20, 2000.

Pursuant to instructions in the February 29, 2000 O.G., we hereby request that the errors which are identified above be corrected in the captioned application to which this request for correction is directed. It is understood that the Patent Office will issue an automatically-generated, corrected Filing Receipt in this and, if applicable, any other affected applications.

Respectfully submitted,

HAMILTON, BROOK, SMITH & REYNOLDS, P.C.

By Lisa M. Treannie
Lisa M. Treannie
Registration No.: 41,368
Telephone: (781) 861-6240
Facsimile: (781) 861-9540

Lexington, MA 02421-4799

Date: 4/5/01



UNITED STATES PATENT AND TRADEMARK OFFICE

COMMISSIONER FOR PATENTS
UNITED STATES PATENT AND TRADEMARK OFFICE
WASHINGTON, D.C. 20231
www.uspto.gov

| APPLICATION NUMBER | FILING DATE | GRP ART UNIT | FIL FEE REC'D | ATTY.DOCKET.NO | DRAWINGS | TOT CLAIMS | IND CLAIMS |
|--------------------|-------------|--------------|---------------|----------------|----------|------------|------------|
|--------------------|-------------|--------------|---------------|----------------|----------|------------|------------|

| | | | | | | | |
|------------|------------|------|---|---------------|----|----|----|
| 09/693,205 | 10/20/2000 | 1646 | 0 | 2862-1021-003 | 22 | 39 | 15 |
|------------|------------|------|---|---------------|----|----|----|

2825.1021-003

FILING RECEIPT



OC000000005621416

Doreen M Hogle Esq
HAMILTON BROOK SMITH & REYNOLDS P C
Two Militia Drive
Lexington, MA 02421-4799

Date Mailed: 12/14/2000

Receipt is acknowledged of this nonprovisional Patent Application. It will be considered in its order and you will be notified as to the results of the examination. Be sure to provide the U.S. APPLICATION NUMBER, FILING DATE, NAME OF APPLICANT, and TITLE OF INVENTION when inquiring about this application. Fees transmitted by check or draft are subject to collection. Please verify the accuracy of the data presented on this receipt. If an error is noted on this Filing Receipt, please write to the Office of Initial Patent Examination's Customer Service Center. Please provide a copy of this Filing Receipt with the changes noted thereon. If you received a "Notice to File Missing Parts" for this application, please submit any corrections to this Filing Receipt with your reply to the Notice. When the PTO processes the reply to the Notice, the PTO will generate another Filing Receipt incorporating the requested corrections (if appropriate).

Applicant(s)

Thomas J. Hudson, residence, NOT PROVIDED;
James Engert, residence, NOT PROVIDED;
Andrea Richter, residence, NOT PROVIDED;

Continuing Data as Claimed by Applicant

THIS APPLN CLAIMS BENEFIT OF 60/160,588 10/20/1999

Foreign Applications

If Required, Foreign Filing License Granted 12/14/2000

Title

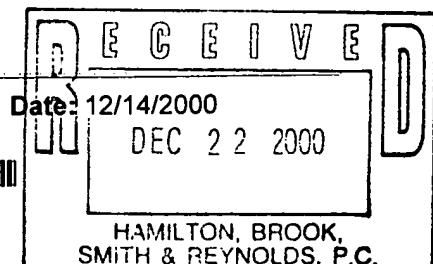
Identification of arsacs mutations and methods of use therefor

Preliminary Class

530

Data entry by : WARREN, EFREM

Team : OIPE



**LICENSE FOR FOREIGN FILING UNDER
Title 35, United States Code, Section 184
Title 37, Code of Federal Regulations, 5.11 & 5.15**

GRANTED

The applicant has been granted a license under 35 U.S.C. 184, if the phrase "IF REQUIRED, FOREIGN FILING LICENSE GRANTED" followed by a date appears on this form. Such licenses are issued in all applications where the conditions for issuance of a license have been met, regardless of whether or not a license may be required as set forth in 37 CFR 5.15. The scope and limitations of this license are set forth in 37 CFR 5.15(a) unless an earlier license has been issued under 37 CFR 5.15(b). The license is subject to revocation upon written notification. The date indicated is the effective date of the license, unless an earlier license of similar scope has been granted under 37 CFR 5.13 or 5.14.

This license is to be retained by the licensee and may be used at any time on or after the effective date thereof unless it is revoked. This license is automatically transferred to any related applications(s) filed under 36 CFR 1.53(d). This license is not retroactive.

The grant of a license does not in any way lessen the responsibility of a licensee for the security of the subject matter as imposed by any Government contract or the provisions of existing laws relating to espionage and the national security or the export of technical data. Licensees should apprise themselves of current regulations especially with respect to certain countries, of other agencies, particularly the Office of Defense Trade Controls, Department of State (with respect to Arms, Munitions and Implements of War (22 CFR 121-128)); the Office of Export Administration, Department of Commerce (15 CFR 370.10 (j)); the Office of Foreign Assets Control, Department of Treasury (31 CFR Parts 500+) and the Department of Energy.

NOT GRANTED

No license under 35 U.S.C. 184 has been granted at this time, if the phrase "IF REQUIRED, FOREIGN FILING LICENSE GRANTED" DOES NOT appear on this form. Applicant may still petition for a license under 37 CFR 5.12, if a license is desired before the expiration of 6 months from the filing date of the application. If 6 months has lapsed from the filing date of this application and the licensee has not received any indication of a secrecy order under 35 U.S.C. 181, the licensee may foreign file the application pursuant to 37 CFR 5.15(b).

PLEASE NOTE the following information about the Filing Receipt:

- The articles such as "a," "an" and "the" are not included as the first words in the title of an application. They are considered to be unnecessary to the understanding of the title.
- The words "new," "improved," "improvements in" or "relating to" are not included as first words in the title of an application because a patent application, by nature, is a new idea or improvement.
- The title may be truncated if it consists of more than 600 characters (letters and spaces combined).
- The docket number allows a maximum of 25 characters.
- If your application was submitted under 37 CFR 1.10, your filing date should be the "date in" found on the Express Mail label. If there is a discrepancy, you should submit a request for a corrected Filing Receipt along with a copy of the Express Mail label showing the "date in."
- The title is recorded in sentence case.

Any corrections that may need to be done to your Filing Receipt should be directed to:

Assistant Commissioner for Patents
Office of Initial Patent Examination
Customer Service Center
Washington, DC 20231

COPY

FILE COPY

-1-

Date: 10/20/00 Express Mail Label No. EL5525 72736 US

Inventors: Thomas J. Hudson, James Engert and Andrea Richter
Attorney's Docket No.: 2825.1021-003

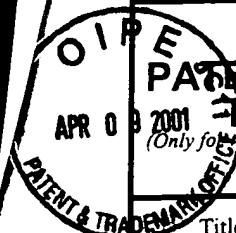
IDENTIFICATION OF ARSACS MUTATIONS AND METHODS OF USE
THEREFOR

RELATED APPLICATIONS

This application claims the benefit of U.S. Provisional application Serial No.
5 60/160,588, filed October 20, 1999, the entire teachings of which are incorporated
herein by reference.

BACKGROUND OF THE INVENTION

Autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) is an early-onset neurodegenerative disease with high prevalence in the Charlevoix-
10 Saguenay-Lac-Saint-Jean (CSLSJ) region of Quebec. Disease progression is rapid through young adulthood, with most patients requiring wheelchairs by their early forties. The disease is characterized by abolished sensory nerve conduction, reduced motor nerve velocity, and a unique clinical feature of hypermyelination of retinal nerve fibers. Additional pathological features include atrophy of the upper cerebellar vermis,
15 absence of Purkinje cells, and possibly abnormal neuronal lipid storage (Bouchard, J-P., *In: Handbook of Clinical Neurology 16: Hereditary neuropathies and spinocerebellar degenerations*, J.M.B.V. de Jong, Ed., pp. 451-459, Elsevier Science Publishers, Amsterdam (1991)). A developmental defect in the myelination of both retinal and peripheral nerve fibers has been proposed as the physiological basis of the disease
20 (Bouchard, J-P., *et al.*, *Neuromuscular Disorders* 8:474-479 (1998)). More than 300

COPY

**UTILITY
PATENT APPLICATION
TRANSMITTAL**
*(Only for new nonprovisional applications under
37 C.F.R. 1.53(b))*

Attorney Docket No: 2825.1021-503

First Named Inventor or Application Identifier Thomas J. Hudson

Express Mail Label No. EL552572736US

FILE COPY

Title of Invention

IDENTIFICATION OF ARSACS MUTATIONS AND METHODS OF USE THEREFOR

APPLICATION ELEMENTS

See MPEP chapter 600 concerning utility patent application contents.

ADDRESS TO: Assistant Commissioner for Patents
Box Patent Application
Washington, D.C. 20231

| | |
|---|--|
| 1. <input type="checkbox"/> Fee Transmittal Form <i>(Submit an original, and a duplicate for fee processing)</i> | 6. <input type="checkbox"/> Microfiche Computer Program (<i>Appendix</i>) |
| 2. <input checked="" type="checkbox"/> Specification [Total Pages [1-57]] <i>(preferred arrangement set forth below)</i> | 7. <input type="checkbox"/> Nucleotide and/or Amino Acid Sequence Submission <i>(if applicable, all necessary)</i> |
| - Descriptive title of the invention | a. <input type="checkbox"/> Computer Readable Copy |
| - Cross References to Related Applications | b. <input type="checkbox"/> Paper Copy (identical to computer copy) |
| - Statement Regarding Fed sponsored R & D | [] Pages |
| - Reference to microfiche Appendix | c. <input type="checkbox"/> Statement verifying identity of above copies |
| - Background of the Invention | |
| - Summary of the Invention | |
| - Brief Description of the Drawings | |
| - Detailed Description | |
| - Claim(s) | |
| - Abstract of the Disclosure | |
| 3. <input checked="" type="checkbox"/> Drawing(s) (35 U.S.C. 113) [Total Sheets [1-22]] [X] Formal [] Informal | 8. <input type="checkbox"/> Assignment Papers (cover sheet & documents) |
| 4. <input type="checkbox"/> Oath or Declaration/POA [Total Pages []] | 9. <input type="checkbox"/> 37 C.F.R. 3.73(b) Statement <input type="checkbox"/> Power of Attorney <i>(when there is an assignee)</i> |
| a. <input type="checkbox"/> Newly executed (original or copy) | 10. <input type="checkbox"/> English Translation Document (<i>if applicable</i>) |
| b. <input type="checkbox"/> Copy from a prior application (37 C.F.R. 1.63(d)) <i>(for continuation/divisional with Box 17 completed)</i> [NOTE Box 5 below] | 11. <input type="checkbox"/> Information Disclosure Statement (IDS)/PTO-1449 <input type="checkbox"/> Copies of IDS Citations |
| i. <input type="checkbox"/> <u>DELETION OF INVENTOR(S)</u> Signed statement attached deleting inventor(s) named in the prior application, see 37 C.F.R. 1.63(d)(2) and 1.33(b). | 12. <input type="checkbox"/> Preliminary Amendment |
| 5. <input type="checkbox"/> Incorporation By Reference (<i>useable if Box 4b is checked</i>) The entire disclosure of the prior application, from which a copy of the oath or declaration is supplied under Box 4b, is considered as being part of the disclosure of the accompanying application and is hereby incorporated by reference therein. | 13. <input checked="" type="checkbox"/> Return Receipt Postcard (MPEP 503) <i>(Should be specifically itemized)</i> |
| 17. If a CONTINUING APPLICATION, check appropriate box and supply the requisite information: <input type="checkbox"/> Continuation <input type="checkbox"/> Divisional <input type="checkbox"/> Continuation-in-part (CIP) of prior application No.: _____ | 14. <input type="checkbox"/> Small Entity <input type="checkbox"/> Statement filed in prior application, status still proper and desired |
| Prior application information: Examiner: _____ | 15. <input type="checkbox"/> Certified Copy of Priority Document(s) <i>(if foreign priority is claimed)</i> |
| | 16. <input type="checkbox"/> Other: _____ |

ACCOMPANYING APPLICATION PARTS

8. Assignment Papers (cover sheet & documents)
9. 37 C.F.R. 3.73(b) Statement Power of Attorney
(when there is an assignee)
10. English Translation Document (*if applicable*)
11. Information Disclosure Statement (IDS)/PTO-1449 Copies of IDS Citations
12. Preliminary Amendment
13. Return Receipt Postcard (MPEP 503)
(Should be specifically itemized)
14. Small Entity Statement filed in prior application, status still proper and desired
15. Certified Copy of Priority Document(s)
(if foreign priority is claimed)
16. Other: _____

17. If a CONTINUING APPLICATION, check appropriate box and supply the requisite information:

 Continuation Divisional Continuation-in-part (CIP) of prior application No.: _____

Prior application information: Examiner: _____ Group Art Unit: _____

18. CORRESPONDENCE ADDRESS

| | | | | | |
|---------|--|-----------|----------------|----------|----------------|
| NAME | Doreen M. Hogle, Esq. HAMILTON, BROOK, SMITH & REYNOLDS, P.C. | | | | |
| ADDRESS | Two Militia Drive | | | | |
| CITY | Lexington | STATE | MA | ZIP CODE | 02421-4799 |
| COUNTRY | USA | TELEPHONE | (781) 861-6240 | FAX | (781) 861-9540 |

| | | | |
|---------------------------------------|------------------|-------------|------------------|
| Signature | Lisa M. Treannie | Date | October 20, 2000 |
| Submitted by Typed or Printed Name | Lisa M. Treannie | Reg. Number | 41,368 |

#3



UNITED STATES PATENT AND TRADEMARK OFFICE

COMMISSIONER FOR PATENTS
UNITED STATES PATENT AND TRADEMARK OFFICE
WASHINGTON, D.C. 20231
www.uspto.gov

| APPLICATION NUMBER | FILING/RECEIPT DATE | FIRST NAMED APPLICANT | ATTORNEY DOCKET NUMBER |
|--------------------|---------------------|-----------------------|------------------------|
| 09/693,205 | 10/20/2000 | Thomas J. Hudson | 2852.1021-003 |

Doreen M Hogle Esq
HAMILTON BROOK SMITH & REYNOLDS P.C.
Two Militia Drive
Lexington, MA 02421-4799

DOCKETED

FORMALITIES LETTER



OC000000005621417

Date Mailed: 12/14/2000

NOTICE TO FILE MISSING PARTS OF NONPROVISIONAL APPLICATION

FILED UNDER 37 CFR 1.53(b)

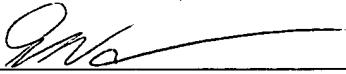
Filing Date Granted

An application number and filing date have been accorded to this application. The item(s) indicated below, however, are missing. Applicant is given TWO MONTHS from the date of this Notice within which to file all required items and pay any fees required below to avoid abandonment. Extensions of time may be obtained by filing a petition accompanied by the extension fee under the provisions of 37 CFR 1.136(a).

- The statutory basic filing fee is missing.
Applicant must submit \$ 710 to complete the basic filing fee and/or file a small entity statement claiming such status (37 CFR 1.27).
- Total additional claim fee(s) for this application is \$1302.
 - \$342 for 19 total claims over 20.
 - \$960 for 12 independent claims over 3 .
- The oath or declaration is missing.
A properly signed oath or declaration in compliance with 37 CFR 1.63, identifying the application by the above Application Number and Filing Date, is required.
- To avoid abandonment, a late filing fee or oath or declaration surcharge as set forth in 37 CFR 1.16(e) of \$130 for a non-small entity, must be submitted with the missing items identified in this letter.
- The balance due by applicant is \$ 2142.**

| | |
|---------|----------|
| 710.00 | 00000024 |
| 1040.00 | 09693205 |
| 342.00 | 00000024 |
| 130.00 | 09693205 |

A copy of this notice MUST be returned with the reply.


Customer Service Center

Initial Patent Examination Division (703) 308-1202

PART 2 - COPY TO BE RETURNED WITH RESPONSE

04/11/2001 JAD001

01 FC:101
02 FC:102
03 FC:103
04 FC:105

